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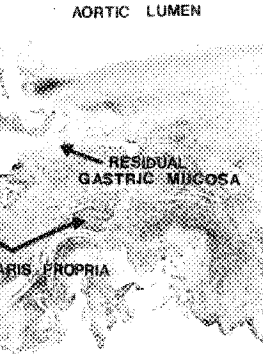

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5, 574–81.

3. GARSTIN W. I. H., JOHNSTON G. W., KENNEDY T. L. & SPENCER E. F. A. (1986) Nissen fundoplication: The unhappy 15%. *J. Royal Coll. Surg. Edin.* **31**, 207–9.
4. CAMPBELL R., KENNEDY T. & JOHNSTON G. W. (1983) Gastric ulceration after Nissen fundoplication. *Brit. J. Surg.* **70**, 406–7.
5. RICHARDSON J. D., LARSON G. M. & POLK H. C. JR (1982) Intrathoracic fundoplication for shortened oesophagus. Treacherous solution to a challenging problem. *Amer. J. Surg.* **143**, 29–35.

Aust. N.Z. J. Surg. 1990, **60**, 825–828

MAZABRAUD'S SYNDROME: INTRAMUSCULAR MYXOMA ASSOCIATED WITH FIBROUS DYSPLASIA OF BONE

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A 53 year old male with a large swelling on the medial aspect of his right thigh was referred with a presumptive diagnosis of soft tissue sarcoma. However, biopsy revealed intramuscular myxoma and X-rays and CT scans suggested fibrous dysplasia of adjacent bone. Angiography had shown an expanded, hypervascular, intramedullary lesion in the femur, and a large avascular soft tissue mass lying medially in the distal thigh. Fibrous dysplasia of the femur was confirmed on bone biopsy. Subsequently one large and two smaller intramuscular myxomata were excised, with an uneventful postoperative course. This case illustrates Mazabraud's syndrome: the rare association between benign intramuscular myxoma and fibrous dysplasia of bone.

Key words: fibrous dysplasia of bone, intramuscular myxoma, Mazabraud's syndrome.

Case report

A plant operator aged 53 years was referred with a provisional diagnosis of soft tissue sarcoma of the right thigh. He gave a 14 year history of progressive enlargement of a swelling on the antero-medial aspect of his distal thigh. Over more recent months the patient had developed pain in the region and had suffered recurrent lateral dislocation of the right patella. His past history and family history were unremarkable. Examination revealed firm, bosselated masses on the medial aspect of the distal thigh (Fig. 1). The largest, which measured 11 × 13 cm, was obviously intramuscular and partially cystic. There were two smaller, firmer masses measuring 4 × 3 cm and 3 × 3 cm which appeared more superficial but nevertheless intramuscular.

X-rays of the right femur showed areas of increased density in the medullary cavity and other areas of ground-glass appearance, consistent with fibrous dysplasia (Fig. 2). An expanded segment in the mid shaft over about 6 cm was noted, below which the medullary cavity had a multiloculated pattern. The adjacent large soft tissue mass was noted. CT scans of the right femur were also suggestive of fibrous dysplasia (Fig. 3). They showed a large intramuscular soft tissue mass, of fluid density and loculated in areas, extending down the medial aspect of the distal one third of the right thigh. Two smaller lesions were seen superiorly and inferiorly. A right femoral arteriogram showed the upper expanded intramedullary lesion to be hypervascular, while the large soft tissue mass lying medially in the distal thigh was avascular. Technetium^{99} bone scan revealed abnormal uptake throughout the right tibia, the mid and proximal portions of the right femur, right humerus, right ulna and right clavicle.

To establish histological diagnoses, needle core biopsies were taken from the soft tissue mass and cylindrical bone biopsies from the right femur and right tibia. Intramuscular myxoma was confirmed and the bone biopsies confirmed the presence of fibrous dysplasia. Two days later the patient suffered a pathological fracture of the mid shaft of the right femur, which was curetted and fixed by insertion of an intramedullary nail. He was discharged

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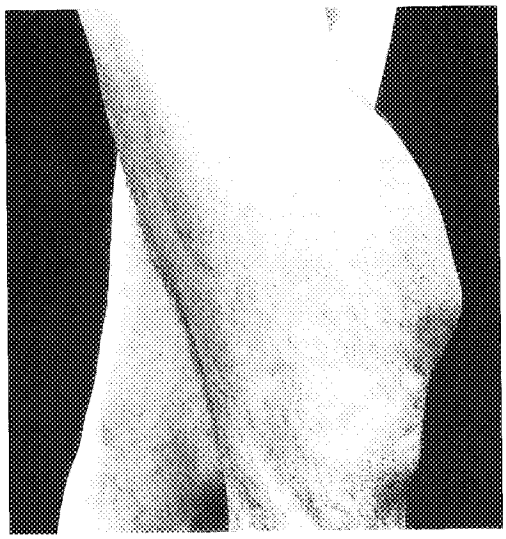

Fig. 1. Preoperative view of right thigh.

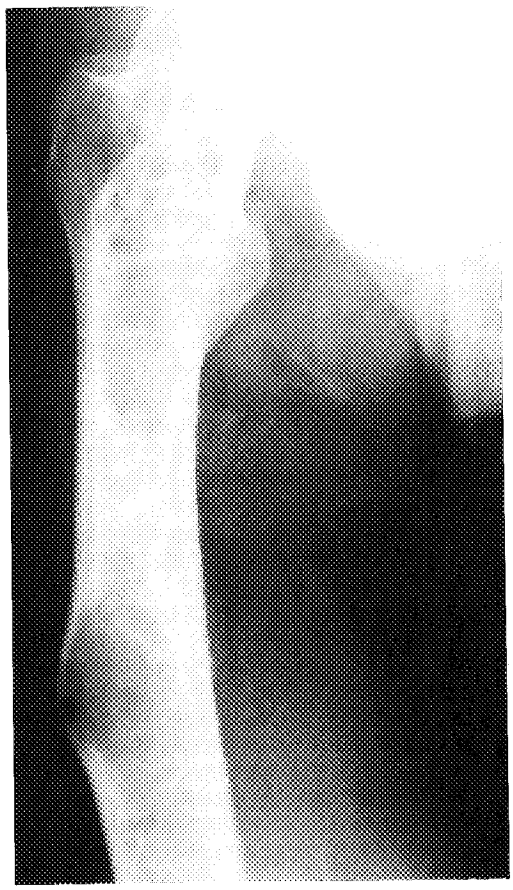

Fig. 2. Anteroposterior radiograph of right femur showing fibrous dysplasia.

after 2 weeks and readmitted 3 months later for removal of the myxomas.

The three tumours were excised locally from the medial aspect of the right thigh under general anaesthesia, with tourniquet control (Fig. 4). The largest tumour was deep to and within vastus medialis and adherent to periosteum medially, whilst the two smaller intramuscular lesions were more superficial. The patient's postoperative course was unremarkable and he was discharged on the seventh postoperative day, able to ambulate with some residual discomfort but no instability. He remains well, with no evidence of residual or recurrent disease, six months later.

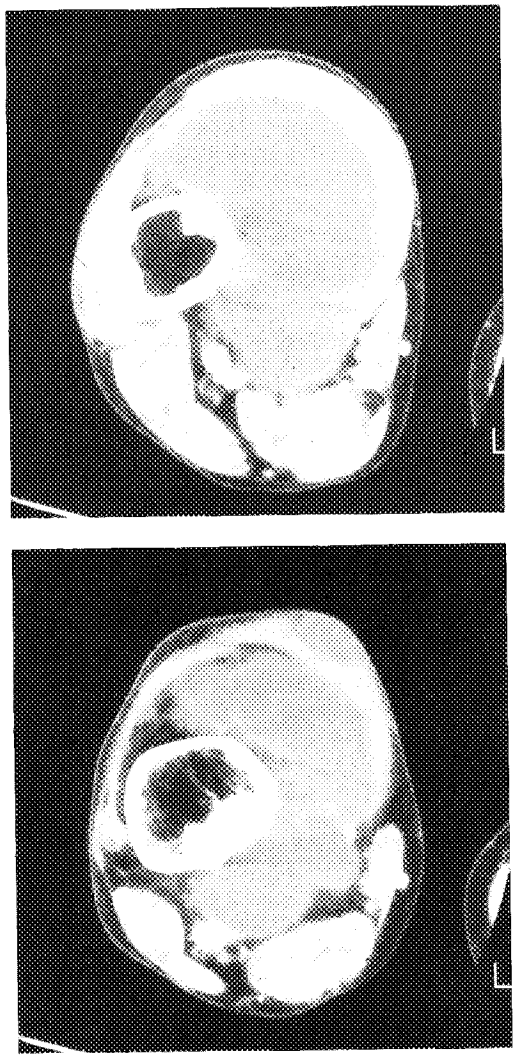

Fig. 3. Computerized axial tomograms of distal right thigh.

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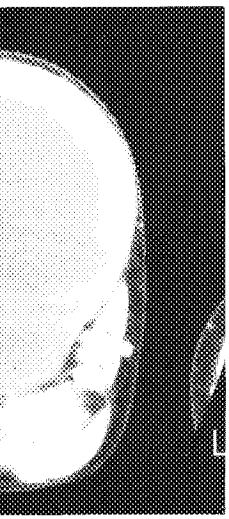

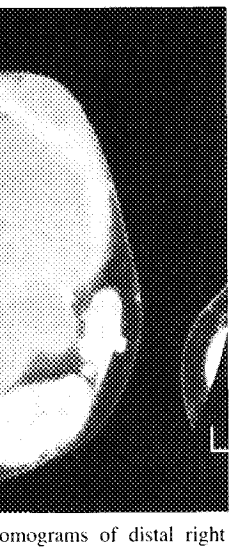

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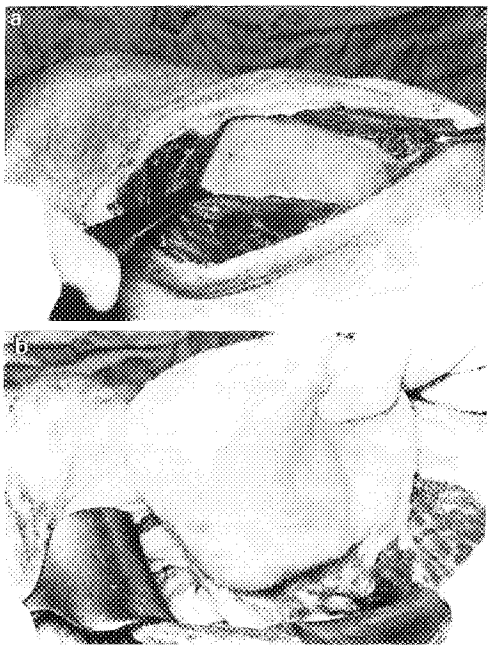

Fig. 4. **(a)** Vastus medialis split and retracted to expose myxoma. **(b)** Myxoma being delivered from thigh.

Discussion

Intramuscular myxoma associated with fibrous dysplasia of bone is a rare syndrome described by Mazabraud in 1957,[1] although coexistence of the two conditions was first observed by Henschen in 1926.[2] In 1928, Krogius reported another case of 'osteitis fibrosa' associated with several myxomas, and similar cases have subsequently been described by others.[3–14] A review of the literature up until 1989 reveals that only 16 cases of the syndrome have been reported altogether.

Intramuscular myxoma is a rare benign tumour of soft tissue thought to arise from fibroblasts that produce excessive amounts of mucopolysaccharides, which in turn inhibit the normal polymerization of collagen. It is a tumour of adult life, occurring primarily between the ages of 40 and 70 years, with a slight female predominance. Presentation is generally nonspecific with a painless palpable mass that is firm, somewhat mobile and often fluctuant. In only 25% of cases is pain or tenderness a feature, although in very large tumours neurovascular symptoms distal to the lesion may be present. Most of the tumours are apparent for many months prior to excision because of the relative lack of symptoms. Sites of predilection for myxomas are the large muscles of the thigh, shoulder, buttock and upper arm, but they also occur in the head and neck region, on the chest wall, in subcutaneous tissue and in cardiac muscle. They vary in size from a few mm to 10 or 15 cm in diameter.

The importance of recognizing these tumours as a separate entity is to distinguish them from richly myxoid malignant tumours, in particular from myxoid liposarcoma, botyroid-type rhabdomyosarcoma, myxoid malignant fibrous histocytoma and myxoid chondrosarcoma. All of these are differentiated by a much greater degree of cellularity, a more pronounced vascular pattern and the presence of specific cellular elements such as lipoblasts, rhabdomyoblasts or chondroblasts. Although the distinction is not always straightforward, it is of great importance because myxomas are benign, with minimal if any tendency to recur, and are cured by simple local excision. There has been no report of malignant degeneration of an intramuscular myxoma.

Fibrous dysplasia is a hamartomatous bony disorder, slightly more frequent in females, which appears in monostotic or polyostotic forms with or without extraskeletal manifestations. These extraskeletal lesions include skin hyperpigmentation, alopecia, hyperthyroidism, diabetes, and renal and cardiovascular malformations.[15] The monostotic form is the most common and the majority of the lesions are in a rib or the craniofacial skeleton, although they also occur in the proximal femur, tibia or humerus. It may be asymptomatic or lead to pathological fracture. Monostotic fibrous dysplasia is usually diagnosed in patients between the ages of 20 and 30 years, and associated skin lesions are infrequent. Polyostotic fibrous dysplasia may be limited to a single limb or to a side of the body, or may occur in a more widespread distribution. Approximately 25% of those suffering the polyostotic form have over half the skeleton involved, and the disease is generally more severe and deforming, with an earlier clinical onset.

The pathology of fibrous dysplasia is essentially identical for all forms of the disease. The normal cancellous bone is replaced by gritty grey-pink rubbery tissue and the endosteal surface is frequently scalloped. Histologically, the areas of fibrous dysplasia consist of compact histiofibroblastic tissue containing thin reticular bone trabeculae not bordered by osteoblasts. Typically, there is an absence of lamellar structure in the bone trabeculae. In adults there tends to be maturation of the dysplastic tissue, decreased numbers of connective tissue cells, maturation of the bone fibroblasts and thickening of the collagenous stroma.[15] Malignant degeneration into sarcoma (osteosarcoma, chondrosarcoma, fibrosarcoma) rarely occurs and in most cases arises in previously irradiated areas. Radiologically the appearance is of a radiolucent area with a well-delineated, smooth or scalloped border associated with focal cortical thinning. The

ground-glass appearance reflects the thin spicules of calcified woven bone. Deformity is common.

The progressive nature of the disease tends to decrease or cease with skeletal maturity. Surgical intervention is indicated when significant deformity or pathological fracture occurs or when significant pain exists. The lesions are adequately treated by curettage and bone grafting; deformities are corrected by osteotomy with internal fixation, and pathological fractures are best treated by curettage and grafting with internal fixation of the fracture.

Where myxoma is associated with fibrous dysplasia of bone it is almost always multiple and in the same anatomical region as the bony changes.[11] This rare association, 'Mazabraud's syndrome', raises the possibility of a localized error in tissue metabolism. The association is more common with the polyostotic form of fibrous dysplasia and, strangely, the myxomata show a predilection for the right side of the body.[11] There is frequently a long interval (up to 20–30 years) between the appearance of fibrous dysplasia, usually noted during the growth period, and the development of the myxomata.[11] The average age of the 16 previously reported cases was 52 years (range 21–72) although the fibrous dysplasia had appeared at a young age in most of the patients. The syndrome was diagnosed late in 80% of cases. Sixty-five per cent of patients exhibited the polyostotic form. In 75% of cases there were multiple myxomata but in most of the remaining 25% full examination of the soft tissues was not performed to exclude further lesions. In none of the patients has recurrence been documented.

In summary, intramuscular myxoma is a rare benign tumour of soft tissue which even more rarely is associated with fibrous dysplasia of bone. Its importance is in its distinction from malignant soft tissue tumours because it can be treated with negligible recurrence rates by simple local excision.

Acknowledgement

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References

1. Mazabraud A. & Girard J. (1957) Un cas particulier de dysplasie fibreuse a localisations osseuses et tendineuses. *Rev. Rhum. Mal. Osteoartic.* **34**, 652–9.
2. Henschen F. (1926) Fall von ostitis fibrosa mit multiplen Tumoren inder Umgebung der Muskulatur. *Verh. Otsch. Ges. Pathol.* **21**, 93–7.
3. Krogius A. (1928) Ein Fall von Ostitis Fibrosa mit miltiplen fibromyxomatoesen Musketumoren. *Acta Chir. Scand.* **64**, 465–71.
4. Vehlinger E. (1940) Osteofibrosis deformans juvenilis *Virchows Arch. Pathol. Anat.* **306**, 255–99.
5. Braunwarth K. (1953) Gleichzeitiges Aufireten von Fibroeser Dysplasie und extraossalen Fibromyxomen. *Fortschr. Geb. Rontgenstr. Nuklearmed.* **78**, 589–94.
6. Laport F., Lefer H. & Malchair G. (1961) Le Syndrome d'Albright. *J. Chir.* **82**, 457–85.
7. Lick R. F. & Viehweger G. (1962) Ein Beitrag zur Diagnose der fibrosen dysplasie des Knochensystems. *Fortschr. Geb. Roentgenstr. Nuklearmed.* **97**, 33–8.
8. Leung T. K. Vauzelle J. L., Patricot L. M., Lejeune E. & Queneau P. (1971) Etude ultrastructurale et cytochemigue d'un myxome musculaire associe a une dysplasie fibreuse. *Ann. Anat. Pathol.* **16**, 417–28.
9. Ireland D. C. R., Soule E. H. & Ivins J. C. (1973) Myxoma of somatic soft tissues. *Mayo Clin. Proc.* **48**, 401–10.
10. Wirth W. A., Leavitt D. & Enzinger F. M. (1976) Multiple intramuscular myxoma. *Cancer* **27**, 1167–73.
11. Mazabraud A., Semat P. & Roze R. (1976) Apropos de l'association de fibromixomes des tissus mous a la dysplasie fibreuse des os. *Presse. Med.* **75**, 2223–8.
12. Berkhoff W. B. C., Ter Bruggen Hugenhaltz F. I. O. & Ingenhoes R. (1981) Causuistische Mededelinger. *Ned. Tijdschr. Geneeska* **125**, 1460–2.
13. Heineman G. & Woerth D. (1983) Zur Osteofibrosis deformans juvenilis. *Langenbecks Arch. Chir.* **197**, 326–36.
14. Sedmak D. D., Hart W. R., Bethodek G. H. & Marks K. H. (1983) Massive intramuscular myxoma associated with fibrous dysplasia of bone. *Clev. Clin. Q.* **50**, 469–72.
15. Biagini R., Ruggieri P., Boriani S. & Picci P. (1987) The Mazabraud Syndrome: Case report and review of the literature. *Ital. J. Orthop. Traumatol.* **13**, 105–11.